Introduction

By 3–4 months of age (when fetal hemoglobin declines to <50% of total), many children with sickle cell anemia (HbSS) and sickle β-thalassemia develop clinically significant hemolytic anemia and impairment of splenic function. In others, although the HbF may remain above 50% these children are still at risk of splenic hypo function. Even though the spleen may be enlarged during the first years of life, its phagocytic function is markedly reduced. Therefore, children with sickle cell anemia are at risk of overwhelming septicemia, often without a primary focus, due to encapsulated organisms, including Streptococcus pneumonia and Haemophilus influenza type B.

This clinical practice guideline has been developed for the management of febrile patients with sickle cell disease who present to the emergency department or are inpatients.

Preventative Management

- To reduce high mortality, we strongly recommend:
  - Early diagnosis of sickle cell anemia by newborn screening and referral to a comprehensive care program for sickle cell disease. With newborn screening in place since November 2006, patients should be seen within 3 months of birth.
  - Prophylactic penicillin or amoxicillin, to be prescribed as soon as sickle cell disease is diagnosed, and continued until at least 5 years of age (to be continued past the age of 5 years in certain circumstances). In patients with significant beta-lactam allergy, trimethoprim-sulfamethoxazole should be used.
  - Vaccination against pneumococcus, meningococcus and haemophilus influenza type B. Annual influenza vaccine is also recommended.
  - Despite these measures, septicemia may still occur. Whenever a child with sickle cell disease has an oral or rectal temperature >38.5°C or an axillary temperature >38°C, he or she should be seen urgently. Febrile young infants (<3 months of age) should have an appropriate infectious work up, irrespective of their sickle cell status.

Clinical Recommendations for Management of Fever in Patients with Sickle Cell Disease

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Fever: Guidelines for Management in Children with Sickle Cell Disease

**Emergency Department Initial Assessment and Management:**

- Complete history, physical exam, and routine/additional investigations using ED Sickle Cell with Fever order set or form.
- Blood culture, CBC, differential, reticulocyte count to be obtained if child presents with cough, chest pain, hypoxia or clinical suspicion for pneumonia/acute chest syndrome.
- Blood type and screen to be obtained if child presents with cough.
- Consult Hematology service for all patients with the following: hemoptysis, instability, epigastric, epigastric, splenic, apical masses, acute abdominal pain, urination, dehydration, or fever; or otherwise concerned about the patient's clinical status.
- Develop a management plan as outlined in Sickle Cell Disease (SCD) emergencies.

**Emergency Department Monitoring:**

- Monitor vital signs.
- Admit child to hospital if: oral or rectal temperature > 38.5ºC or axillary °C or axillary °C or otherwise concerned.
- Refer to Pediatric Emergency Department (PED) for admission to hospital if: child is afebrile but presents with persistent hypoxia.

**Inpatient Care:**

- Refer to Inpatient Management document.
- Refer to Communication of Patient Information During Hospital Transfer policy.
- Consult Hematology service if: child presents with cough, chest pain, hypoxia, or otherwise concerned about the patient's clinical status.
- Consult Pediatric Medicine for admission to hospital.
- Complete history, physical exam, and investigations.

**Discharge home from ED with appropriate follow up:**

- Refer to Discharge Management document.
- Child discharged home from ED with appropriate follow up.

**Discharge home from PED with appropriate follow up:**

- Child discharged home from PED with appropriate follow up.
Fever: Guidelines for Management in Children with Sickle Cell Disease

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Fever: Guidelines for Management in Children with Sickle Cell Disease

References


Related documents

- Acute Painful Episodes Vaso-occlusive Crisis: Guidelines for Management in Children with Sickle Cell Disease
- Acute Chest Syndrome or Pneumonia: Guidelines for Management in Children with Sickle Cell Disease

Attachments:

SCD fever_discharge planning process.pdf

SCD fever_inpatient management.pdf
### Fever: Guidelines for Management in Children with Sickle Cell Disease

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**Version:** 4

- [SCD pain plan_july 2015.pdf](#)
- [SCD fever_out patient follow up.pdf](#)
- [SCD fever_criteria for admission.pdf](#)
- [care pathway_final_IPPCC approved April 2019.pdf](#)
- [Revision History.docx](#)
- [SC_clinic follow up.pdf](#)

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